PART XVI Rheumatic Diseases of Childhood

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1. Nonsteroidal anti-inflammatory drugs (NSAIDs) are prescribed to decrease acute and chronic inflammation associated with various rheumatic diseases; however, many adverse effects have been encountered with long term use.
Of the following, the NSAID that has the highest toxicity is

A. celecoxib
B. meloxicam
C. indomethacin
D. naproxen
E. ibuprofen

2. A 7-year-old boy developed small hypopigmented depressed scars after fingernail scratches; he has been treated with nonsteroidal antiinflammatory drugs (NSAIDs) for a rheumatic disease 6 weeks ago.
Of the following, the NSAID that is **MOST** likely to cause such a unique skin reaction is

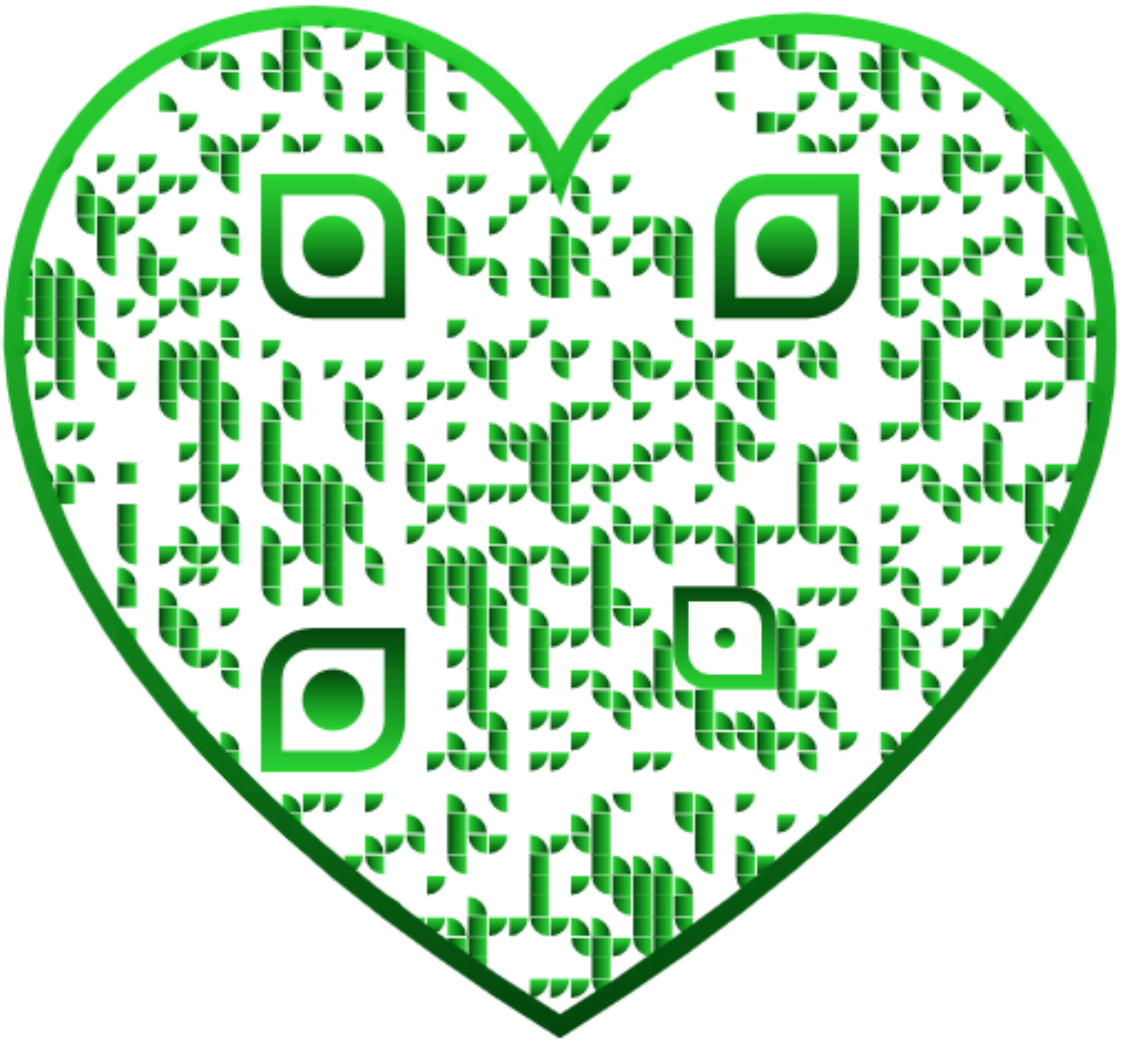
A. celecoxib
B. meloxicam
C. indomethacin
D. naproxen
E. ibuprofen

3. Hydroxychloroquine sulfate is an antimalarial drug important in the treatment of SLE and dermatomyositis, particularly cutaneous manifestations of disease and to reduce lupus flares.
Of the following, the **MOST** important procedure that should be done routinely during the course of administration is

A. gastric endoscopy
B. bone marrow examination
C. muscle biopsy
D. glucose-6-phophate dehydrogenase enzyme level assay
E. ophthalmological examination

4. Sulfasalazine is an effective drug in many rheumatic diseases; however, it is associated with severe systemic hypersensitivity reaction.
It is approved in all the following rheumatic disease **EXCEPT**

A. polyarticular juvenile idiopathic arthritis (JIA)
B. oligoarticular JIA



C. systemic JIA
D. peripheral arthritis associated with juvenile ankylosing spondylitis
E. enthesitis associated with juvenile ankylosing spondylitis

5. You are meeting parents of a 14-year-old girl who has been treated with a monthly intravenous cyclophosphamide for SLE-associated renal failure for the last 6 months; the mother is asking about the long-term complications of this drug.
All the following are long-term complications **EXCEPT**

A. bone marrow suppression
B. bladder cancer
C. leukemia
D. lymphoma
E. infertility

6. Juvenile idiopathic arthritis has many subtypes. Arthritis must be present to make a diagnosis of any subtype; involved joints oftenly have the following signs **EXCEPT**

A. swelling
B. warm
C. erythema
D. limitation of movement
E. pain on movement

7. Oligoarthritis, the most common subtype of juvenile idiopathic arthritis, is defined as involving ≤4 joints within the 1st 6 mo of disease onset.
Of the following, the **MOST** commonly affected joint is

A. hip
B. knee
C. elbow
D. wrist
E. metatarsal

8. Oligoarthritis predominantly affects the joints of the lower extremities rather than upper extremity joints. Which of the following joints is never a presenting sign of oligoarthritis?

A. hip
B. knee
C. ankle
D. metatarsal
E. interphalangeal

9. A 4-year-old girl recently diagnosed with persistent oligoarticular juvenile idiopathic rheumatoid arthritis (JIA); she has 3 involved joints including the right knee, right ankle, and left elbow; antinuclear antigen (ANA) is significantly positive.
Of the following, the **MOST** important step in the management of this girl is

A. regular examination of locomotor system
B. periodic slit-lamp examination
C. periodic ANA monitoring
D. periodic erythrocyte sedimentation rate (ESR) monitoring
E. frequent C-reactive protein (CRP) monitoring

10. Antinuclear antigen (ANA) measurement test is useful in some rheumatologic diseases especially with persistent oligoarticular juvenile idiopathic rheumatoid arthritis (JIA)**.**
All the following are more likely to be correlated with ANA positivity **EXCEPT**

A. anterior uveitis
B. younger age at disease onset
C. female sex
D. symmetrical arthritis
E. lower number of involved joints over time

11. Rheumatoid factor (RF)–positive polyarthritis is characterized by aggressive symmetric inflammation of joints of both upper and lower extremities.
Of the following, the extra articular manifestation that is almost exclusively occur in RF-positive individuals is

A. fever
B. evanescent rash
C. uveitis
D. extensor surfaces nodules
E. pericarditis

12. A 5-year-old boy develops acute onset of high spiking fevers, lymphadenopathy, hepatosplenomegaly, and purpura; he has been diagnosed with systemic Juvenile idiopathic arthritis (JIA) since early childhood treated with antiinflammatory drugs. You suspect macrophage activating syndrome (MAS).
Of the following, the **BEST** test that distinguishes MAS from a flare of the primary disease is

A. leucopenia
B. falling ESR
C. hypofibrinogenemia
D. hypertriglyceridemia
E. evidence of hemophagocytosis in the bone marrow

13. Rheumatoid factor (RF)–positive polyarthritis usually accounts for < 10% of all juvenile idiopathic arthritis (JIA) cases. The articular manifestation pattern is characterized by involvement of ≥5 joints in both upper and lower extremities.
Of the following, the **LEAST** effective drug to induce remission for this subtype of JIA is

A. NSAIDs
B. methotrexate
C. TNF-α antagonists
D. IL-1 inhibitors
E. IL-6 inhibitors

14. A 5-year-old boy has systemic juvenile idiopathic arthritis (sJIA) with systemic manifestations including fever, hepatosplenomegaly, lymphadenopathy, and pericarditis.
Of the following, the **BEST** initial treatment for this boy is

A. NSAIDs
B. systemic glucocorticoids
C. TNF-α inhibitors
D. IL-1 antagonists
E. IL-6 antagonists

15. You are discussing with the medical students the role of systemic steroids for the management of rheumatologic diseases.
Your discussion should include all the following statements **EXCEPT**

A. they are recommended for management of severe systemic illness
B. they are used for bridge therapy during the wait for therapeutic response to a disease modified anti rheumatic drugs (DMARD)
C. they are effective for control of uveitis
D. they prevent joint destruction
E. they impose risks of severe toxicities

16. Chronic uveitis is one of the extraarticular manifestations of various subtypes of juvenile idiopathic arthritis.
All the following are risk factors for the development of uveitis **EXCEPT**

A. oligoarthritis subtype
B. female gender
C. ANA-positivity
D. onset of arthritis earlier than 6 yr of age
E. severity of arthritis

17. The child with polyarticular JIA often has a more prolonged course of active joint inflammation and requires early and aggressive therapy.
Of the following, the predictor that carries the **WORST** prognosis is

A. old age at onset
B. rheumatoid factor (RF) seronegativity
C. absence of rheumatoid nodules
D. small numbers of affected joints
E. hip joint involvement

18. Spondyloarthritis may be overlapped clinically with other forms of juvenile idiopathic arthritis (JIA). Clinical manifestations that help to distinguish spondyloarthritis from other forms of juvenile arthritis include all the following **EXCEPT**

A. arthritis of the sacroiliac joints
B. arthritis of the hips
C. enthesitis
D. asymptomatic uveitis
E. gastrointestinal inflammation

19. Enthesitis-Related Arthritis (ERA) is a subtype of rheumatic disease that falls in the juvenile idiopathic arthritis (JIA) category.
Of the following, the clinical feature that is highly suggestive of ERA is

A. symmetrical arthritis early in the disease course
B. early involvement of more than 5 joints
C. involvement of upper limb joints
D. Inflammation of the small joints of the foot
E. asymmetrical enthesitis

20. Spondyloarthritides are complex diseases in which susceptibility is largely genetically determined. Which of the following characteristics is never associated with such diseases?

A. enthesitis
B. peripheral arthritis
C. axial arthritis
D. HLA-B27 positivity
E. rheumatoid factor positivity

21. You are discussing with medical students Juvenile Ankylosing Spondylitis (JAS) subtype of spondyloarthritides. One of the students asks you about the features that distinguish this disease from the adult-onset ankylosing spondylitis (AOAS).
All the following are true regarding JAS in comparison with AOAS **EXCEPT**

A. it is present in patients <16 yr old
B. axial disease occurs more frequently early in the disease course
C. inflammatory back pain is less frequent at disease onset
D. enthesitis occurs more commonly
E. peripheral arthritis is more common

22. Reactive arthritis is defined as joint inflammation caused by a sterile inflammatory reaction following a recent infection.
Of the following, the **LEAST** likely micro-organism that may cause reactive arthritis is

A. Clostridium difficile
B. Salmonella paratyphi
C. Shigella flexneri
D. Yersinia enterocolitica
E. Campylobacter jejuni

23. The distinction between postinfectious arthritis and reactive arthritis is not always clear. All the following are common features of postinfectious arthritis, rather than reactive arthritis **EXCEPT**

A. there is a transient joint swelling or pain
B. it lasts less than 6 wk
C. chronic spondyloarthritis is a common sequelae
D. it may be encountered after group A streptococcus infection
E. rubella infection is a common causative agent

24. You are meeting parents of 2-year-old girl who is going to receive rubella vaccine; the mother is concerned regarding arthritis that may develop after immunization.
Of the following, the statement that should be included during the discussion is

A. it is usually frequently encountered after immunization
B. it typically affects the large joints of the upper extremity
C. it is more common in girls
D. it is uncommon in children
E. arthralgia usually begins immediately after administration of immunization

25. Postinfectious arthritis describes arthritis that occurs after infectious illnesses, mainly viruses.
Of the following, the **LEAST** likely causative viral infection is

A. rubella
B. varicella-zoster
C. cytomegalovirus
D. Epstein-Barr virus
E. herpes-simplex virus

26. A 4-year-old boy has an acute severe left groin pain that refers to thigh. Examination reveals limitation of movement of the left hip joint. Lab investigations reveal normal blood count and indices, and normal ESR; radiologic imaging shows widening of the hip joint space with effusion. You suspect transient synovitis.
Of the following, the **BEST** therapeutic approach for this boy is

A. systemic steroids

B. methotrexate
C. aspiration of joint fluid
D. intravenous antibiotics
E. observation

27. Reactive arthritis has the potential for evolving to chronic arthritis, especially after bacterial enteric infection or genitourinary tract infection (UTI).
Of the following, the microorganism that is **MOST** likely causing chronic arthritis after UTI is

A. Escherichia coli
B. Chlamydia trachomatis
C. Proteus mirabilis
D. Pseudomonas aeruginosa
E. Staphylococcus saprophyticus

28. Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating autoantibodies directed against self-antigens. It occurs in both children and adults.
All the following are features of childhood SLE **EXCEPT**

A. it is has a more severe course
B. there is more widespread organ involvement
C. it usually presents before 5 year of age
D. fever, fatigue, hematologic abnormalities, and arthritis are common clinical manifestations
E. although renal disease is asymptomatic, it is oftenly present as nephrotic syndrome in adolescent age group

29. Skin is a commonly involved organ by SLE. There are different cutaneous manifestations.
Of the following, the skin manifestation that is **MOST** suggestive of SLE in children is

A. malar rash
B. discoid rash
C. photosensitive rash
D. cutaneous vasculitis
E. livedo reticularis

30. Drug-induced lupus refers to the presence of SLE manifestations triggered by exposure to specific medications, including antibiotics.
Of the following, the drug that is **MOST** likely associated with drug-induced lupus is

A. isoniazid
B. rifampin
C. nitrofurantoin

D. penicillin
E. tetracycline

31. Systemic lupus erythematosus (SLE) is often characterized by periods of flare and disease quiescence or may follow a more smoldering disease course.
All the following lab tests correlate with active disease **EXCEPT**

A. positive anti-nuclear antibody titer
B. positive anti–double-stranded DNA level
C. low serum complement level
D. high erythrocyte sedimentation rate
E. elevated C-reactive protein (CRP) value

32. A 12-year-old female adolescent has a recent diagnosis of SLE; she has a faint malar rash and mild arthritis including both elbow and knee joints as well as the joints of small fingers.
Of the following, the **MOST** appropriate initial therapy for this patient is

A. systemic steroids
B. methotrexate
C. leflunomide
D. cyclophosphamide
E. hydroxychloroquine

33. A 3-week-old male baby develops malar rash involving the face and periorbital area after exposure to sun light; you suspect neonatal lupus.
Of the following, the **NEXT** step of management is

A. measurement of anti-Ro and anti-La antibodies of the mother
B. measurement of anti-Ro and anti-La antibodies of the baby
C. cardiac evaluation of the mother
D. cardiac evaluation of the baby
E. hematological evaluation of the baby

34. You are meeting a pregnant mother who has had a previous baby with congenital heart block due to neonatal lupus; she has a positive anti-SSA and anti-SSB antibodies. All the following are therapeutic modalities to prevent occurrence of congenital fetal cardiac complications **EXCEPT**

A. fluorinated corticosteroids
B. intravenous immunoglobulin
C. plasmapheresis
D. hydroxychloroquine
E. methotrexate

35. Juvenile dermatomyositis (JDM) is the most common inflammatory myositis in children characterized by skin rash and proximal muscle weakness.
All the following are common cutaneous manifestations of JDM **EXCEPT**

A. heliotrope rash of the eyelids
B. photosensitivity to ultraviolet light
C. facial erythema sparing the nasolabial folds
D. Gottron papules
E. periungual telangiectasias

36. Treatment of juvenile dermatomyositis (JDM) includes pharmacological and non-pharmacological therapy. The mainstay of pharmacological therapy is corticosteroids that should be integrated with non-pharmacological therapy.
All the following are recommended as part of non-pharmacological treatment program in JDM **EXCEPT**

A. physical therapy
B. occupational therapy
C. avoidance of sun exposure
D. bed rest
E. social and psychological support

37. Most complications from Juvenile dermatomyositis (JDM) are related to prolonged and severe weakness; secondary complications from medical treatments are also common.
Of the following, the **LEAST** likely recognized complications of JDM is

A. aspiration pneumonia
B. gastrointestinal bleeding
C. cardiac arrhythmias
D. muscle atrophy
E. skin calcifications

38. Juvenile localized scleroderma (JLS) is generally insidious with skin manifestations that vary according to disease subtype. Up to 25% of children with LS have extracutaneous manifestations, including arthritis and neurological symptoms.
Of the following, the subtype that is **MOST** commonly associated with neurological manifestations is

A. plaque morphea
B. eosinophilic fasciitis
C. generalized morphea
D. en coup de sabre
E. morphea profunda

39. Juvenile systemic scleroderma (JSS) has a prolonged course with periods of remission and exacerbation. It is characterized by multisystem organ involvement including skin, locomotor system, CNS, and viscera.
Of the following, the **MOST** common early cutaneous manifestation is

A. edema of the dorsum of the hands and fingers
B. induration and fibrosis of the skin
C. flexion contractures at the elbows, hips, and knees
D. skin ulceration over pressure points
E. subcutaneous calcifications

40. You are discussing Raynaud phenomenon (RP) and Raynaud disease (RD) with medical students. You state that RP is usually associated with rheumatic diseases, while RD is independent of an underlying rheumatic disease.
Of the following, the feature that is **MOST** likely consistent with RD, rather than RP is

A. presence in early childhood
B. absence of tissue necrosis and gangrene
C. asymmetric occurrence
D. presence of periungual telangiectasia
E. associated pain and paresthesia

41. Behçet disease (BD) is a primary variable vessel vasculitis, characterized by exacerbations and remissions. The hallmark of the disease is oral ulceration.
All the following are characteristic features of the oral ulcers **EXCEPT**

A. very painful
B. recurrent
C. involve any location in the oral cavity
D. heal with scarring
E. last 3-10 days

42. You are evaluating a 16-year-old female adolescent who has been diagnosed with Behçet disease since the age of 12 year; she recently develops oral genital ulcers; examination reveals multiple oral ulcers involving the hard palate, multiple genital ulcers involving the labia majora, and erythema nodosum. There is no major organ involvement and uveitis.
Of the following, the **BEST** treatment for this patient is

A. colchicine
B. azathioprine
C. cyclophosphamide
D. systemic steroids
E. interferon alpha

43. Primary Sjögren syndrome, although rare, may occur in children between 9-10 year.

Of the following, the **MOST** common manifestation in children is

A. recurrent parotitis
B. sicca symptoms
C. polyarthritis
D. vulvovaginitis
E. hepatitis

44. Familial Mediterranean fever (FMF) is a recessively inherited autoinflammatory disease usually characterized by recurrent self-limited episodes of fever, serositis, arthritis, and skin rash.
Of the following, the hallmark cutaneous finding of this disease is

A. erysipelas-like erythema overlying the dorsum of the foot
B. morbilliform rash
C. migratory rash overlying area of myalgia
D. cold-induced urticaria like lesions
E. generalized pustulosis

45. Amyloidosis is the most serious complication of familial Mediterranean fever (FMF), and in its absence FMF patients may live a normal life span.
Of the following, the organ that is not affected by secondary amyloidosis of FMF is

A. kidney
B. lung
C. nerve
D. heart
E. teste

46. Amyloidosis is a disease caused by protein misfolding; the type referred as amyloid A (AA), amyloidosis usually develops in patients with chronic inflammatory states, especially juvenile idiopathic arthritis.
In which of the following subtypes of JIA there is a highest prevalence of AA amyloidosis?

A. systemic JIA
B. oligoarticular JIA
C. polyarticular JIA
D. psoriatic arthritis
E. enthesitis-related arthritis

47. Although there is no established therapy to AA amyloidosis, colchicine may be an effective drug to prevent the development of amyloidosis.
Of the following, the disease that is respond to colchicine in preventing AA amyloidosis is

A. juvenile idiopathic arthritis

B. ankylosing spondylitis
C. familial Mediterranean fever
D. hyper IgD syndrome
E. cryopyrin-associated periodic syndrome

48. Kawasaki disease (KD) is an acute febrile illness of childhood characterized by vasculitis with a predilection for the coronary arteries.
Predictors of poor outcome across several studies include all the following **EXCEPT**

A. old age
B. male gender
C. persistent fever
D. poor response to IVIG
E. elevated C-reactive protein levels

49. Kawasaki disease (KD) has unique clinical manifestations; however, less consistent clinical presentation may occur.
All the following are common classic clinical features of KD **EXCEPT**

A. bilateral non exudative bulbar conjunctivitis
B. cracked lips
C. edema and erythema of the hands and feet
D. vesicular rash
E. unilateral non suppurative cervical lymphadenopathy

50. You are evaluating a 1-year-old boy with an established diagnosis of Kawasaki disease (KD) since 4 days; the caring nurse asks you about cardiac involvement during this stage.
All the following are possible cardiac complications at this stage **EXCEPT**

A. myocarditis
B. pericarditis
C. coronary artery aneurysm
D. mitral regurgitation
E. cardiogenic shock

51. A follow up two-dimensional echocardiography is performed to a 1.5-year-old boy with Kawasaki disease (KD) 3 weeks after the diagnosis; it shows a small solitary aneurysm of the left anterior descending coronary artery without thrombosis.
Of the following, the **MOST** appropriate therapy for this child is

A. aspirin for 6 months
B. life-long aspirin
C. IVIG and aspirin for 14 days
D. aspirin and clopidogrel antiplatelet for 8 weeks
E. aspirin and warfarin for 6 months

52. Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood affecting small vessels in the skin, joints, gastrointestinal tract, and kidney. Gastrointestinal involvements occur in up to 80% of children with HSP.
Of the following, the **LEAST** gastrointestinal manifestation that may occur in children with HSP is

A. abdominal pain
B. vomiting and diarrhea
C. paralytic ileus
D. melena
E. intussusception

53. A 5-year-old boy has acute onset of abdominal pain and melena; he has been diagnosed with Henoch-Schönlein purpura (HSP) one week before. Examination reveals symmetrical palpable purpura overlying the lower extremities and buttock; other examinations are unremarkable.
Of the following, the **BEST** treatment for this boy is

A. supportive measures
B. steroids
C. azathioprine
D. cyclophosphamide
E. mycophenolate mofetil

54. Renal disease is the major long-term complication, occurring in 1-2% of children with Henoch-Schönlein purpura (HSP). Chronic HSP renal disease is managed with a variety of immunosuppressants.
Of the following, the **LEAST** effective drug for the treatment of renal disease in HSP is

A. prednisolone
B. azathioprine
C. cyclophosphamide
D. cyclosporine
E. mycophenolate mofetil

55. Childhood vasculitis encompasses a broad spectrum of diseases that share in common inflammation of the blood vessels. Vascular injury includes small, medium, and large vessels.
Of the following, the disease that affects predominantly large blood vessels is

A. Henoch-Schönlein purpura
B. granulomatosis with polyangiitis (wegener granulomatosis)
C. childhood polyarteritis nodosa
D. Kawasaki disease
E. Takayasu arteritis

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ANSWERS
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1.**(C).** Certain agents (indomethacin) have a higher risk of toxicity than others (ibuprofen); naproxen has an intermediate risk. Selective COX-2 inhibitors (such as celecoxib and meloxicam) inhibit receptors responsible for promoting inflammation with potential for fewer gastrointestinal adverse effects.
2.**(D).** Naproxen is more likely than other NSAIDs to cause pseudoporphyria. Pseudoporphyria is more likely to occur in fair-skinned individuals and on sun-exposed areas. If pseudoporphyria develops; the inciting NSAID should be discontinued because scars can persist for years or be permanent.
3.**(E).** The most significant potential adverse effect is retinal toxicity, which occurs rarely but results in irreversible color blindness or loss of central vision. Complete ophthalmologic examinations, including assessment of peripheral vision and color fields, are conducted at baseline and every 6-12 mo to screen for retinal toxicity.
4.**(C).** Sulfasalazine is generally considered contraindicated in children with active systemic JIA because of increased hypersensitivity reactions.
5.**(A).** Nausea, vomiting, anorexia, alopecia, mucositis, hemorrhagic cystitis, and bone marrow suppression are potential short-term adverse effects.
6.**(C).** Involved joints are often swollen, warm to touch, and painful on movement or palpation with reduced range of motion, but usually are not erythematous.
7.**(B).** In oligoarthritis, it predominantly affects the large joints of the lower extremities, such as the knees and ankles. Isolated involvement of upper extremity large joints is less common.
8.**(A).** Isolated involvement of the hip is almost never a presenting sign and suggests extended oligoarticular juvenile idiopathic arthritis.
9.**(B).** The presence of a positive ANA confers increased risk for asymptomatic anterior uveitis, requiring periodic slit-lamp examination.
10.**(D).** ANA positivity is more likely to be correlated with asymmetric arthritis.
11.**(D).** Rheumatoid nodules on the extensor surfaces of the elbows, spine, and over the Achilles tendons, although unusual, are associated with a more severe course and almost exclusively occur in RF-positive individuals.
12.**(E).** The most important indicators of MAS include a falling platelet count, extreme hyperferritinemia, increased liver enzymes, falling leukocyte count, persistent, continuous fever ≥38°C, falling ESR, hypofibrinogenemia, and hypertriglyceridemia. The diagnosis is confirmed by bone marrow biopsy demonstrating hemophagocytosis.
13.**(A).** NSAIDs alone rarely induces remission in children with polyarthritis or systemic JIA. Methotrexate is the oldest and least toxic effective drug. Biologic medications that

inhibit proinflammatory cytokines, such as TNF-α, IL-1, and IL-6, demonstrated excellent disease control.

14.**(B).** TNF inhibition is not as effective for the systemic symptoms found in sJIA. When systemic symptoms dominate systemic steroids are started followed by the initiation of IL-1 or IL-6 antagonist therapy, which often induces a dramatic and rapid response.

15.**(D).** Systemic steroids do not prevent joint destruction.

16.**(E).** There is no association between the activity or severity of arthritis and uveitis.

17.**(E).** Disease involving the hip and hand and wrist is associated with a poorer prognosis and may lead to significant functional impairment. All other mentioned factors carry a good outcome.

18.**(D).** Symptomatic, rather than asymptomatic, eye inflammation (acute anterior uveitis) is a clinical feature suggestive of spondyloarthritis.

19.**(D).** In ERA, the arthritis is typically asymmetric and involves 4 or fewer joints during the 1st 6mo of the disease. The most frequently affected joints are the knees, ankles, and hips. Enthesitis is typically symmetric and most commonly affects the lower limbs. Inflammation of the small joints of the foot, or tarsitis, is highly suggestive of ERA.

20.**(E).** Positive rheumatoid factor is never associated with all subtypes of spondyloarthritides.

21.**(B).** In comparison to adult-onset AS, axial disease and inflammatory back pain are less frequent at disease onset, while enthesitis and peripheral arthritis is more common.

22.**(A).** Reactive arthritis typically follows enteric infection with Salmonella sp., Shigella flexneri, Yersinia enterocolitica, Campylobacter jejuni, orgenitourinary tract infection with Chlamydia trachomatis. Escherichiacoli and Clostridium difficile are also causative enteric agents, although less common.

23.**(C).** Postinfectious arthritis does not necessarily share the typical spondyloarthritis pattern of joint involvement.

24.**(D).** Rubella-associated arthropathy may follow natural rubella infection and, infrequently, rubella immunization. It typically occurs in young women, with an increased frequency with advancing age, and is uncommon in preadolescent children and in males. Arthralgia of the knees and hands usually begins within 7 days of onset of the rash or 10-28 days after immunization.

25.**(D).**

26.**(C).** In transient synovitis (toxic synovitis), aspiration of joint fluid is often necessary to exclude septic arthritis and typically results in dramatic clinical improvement.

27.**(B).**

28.**(C).** Childhood SLE is rare before 5 yr of age.

29.**(B).**

30.**(A).**

31.**(A).** ANA titers are not reflective of disease activity; therefore, repeating ANA titers is not helpful in disease management.

32.**(E).** Hydroxychloroquine is recommended for all individuals with SLE if tolerated. In addition to treating mild SLE manifestations such as rash and mild arthritis, hydroxychloroquine prevents SLE flares, improves lipid profiles, and may have a beneficial impact on mortality and renal outcomes. Corticosteroids are a mainstay for treatment of significant manifestations of SLE. Methotrexate, leflunomide, and azathioprine are often used to treat persistent moderate disease, including arthritis, significant cutaneous or hematologic involvement, and pleural disease. Cyclophosphamide is reserved for the most severe, potentially life threatening SLE manifestations, such as renal, neurologic, and cardiopulmonary disease.
33.**(D).** The most serious complication due to neonatal lupus is congenital heart block.
34.**(E).**
35.**(C).** In JDM, facial erythema is crossing the nasolabial folds, in contrast to the malar rash without nasolabial involvement typical of systemic lupus erythematosus.
36.**(D).** Bed rest is not indicated, because weight bearing improves bone density and prevents contractures.
37.**(C).** Cardiac involvement by JDM is rare.
38.**(D).** Children with en coup de sabre may have symptoms unique to central nervous system involvement, such as seizures, hemifacial atrophy, ipsilateral uveitis, and learning/behavioral changes.
39.**(A).**
40.**(B).** RD often begins in adolescence and is characterized by symmetric occurrence, the absence of tissue necrosis and gangrene, and the lack of manifestations of an underlying rheumatic disease. Children have normal nail-fold capillaries. Pain and paresthesia indicate ischemic changes, associated only with RP due to rheumatic diseases.
41.**(D).** The oral ulcers heal without scarring.In contrast; the genital ulcers heal with scars.
42.**(A).** In patients without major organ involvement, colchicine significantly improves oral and genital ulcers, skin features, and disease activity.In pediatric patients with vascular involvement with venous thrombosis, steroids and azathioprine have been used, whereas those with pulmonary arterial or cardiac involvement are initially treated with cyclophosphamide. Interferon alpha can be used for CNS vasculitis.
43.**(A).** Recurrentparotid gland enlargement and parotitis are the most common manifestations in children (>70%), whereas sicca syndrome (dry mouth, painful mucosa, sensitivity to spicy foods, halitosis, widespread dental caries) predominate in adults.
44.**(A).**
45.**(C).** Amyloidosis may develop most commonly in the kidneys, gastrointestinal tract, spleen, lungs, testes, thyroid, and adrenals. Rarely, cardiac amyloidosis may develop; macroglossia and amyloid neuropathy is generally not seen with the amyloidosis of FMF.

46.**(A).** JIA is a rheumatic disease that is associated with the development of AA amyloidosis with the highest prevalence in patients with systemic JIA followed by those with polyarticular disease.
47.**(C).** Unlike AA amyloidosis associated with FMF, AA amyloidosis associated with other autoinflammatory diseases (including TRAPS, cryopyrin-associated periodic syndrome, and HIDS) and chronic rheumatic diseases (JIA, RA, and ankylosing spondylitis) do not respond to colchicine.
48.**(A).** Young age is poor predictor. Other lab findings associated with poor outcome are neutrophilia, thrombocytopenia, transaminitis, hyponatremia, hypoalbuminemia, elevated levels of N-terminal-probrain natriuretic protein.
49.**(D).** Typical rash is maculopapular, erythema multiforme, or scarlatiniform. Bullous, pustular, or vesicular rashes are uncommon skin manifestations.
50.**(C).** Coronary artery aneurysm usually develops in the subacute phase in the 2nd or 3rd week of the illness.
51.**(B).** The patient with KD who has had a small solitary aneurysm should continue aspirin indefinitely. Patients with larger or numerous aneurysms may require the addition of other antiplatelet agents or anticoagulation.
52.**(E).** Intussusception, mesenteric ischemia, and intestinal perforation are uncommon in children with HSP.
53.**(B).** Steroid is most often used to treat significant gastrointestinal involvement or other life-threatening manifestations.
54.**(A).**
55.**(E).**